Early results of single-stage biventricular repair of severe aortic hypoplasia or atresia with ventricular septal defect and normal left ventricle

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Objective: Biventricular repair of aortic atresia (or severe aortic hypoplasia) is possible in the presence of a ventricular septal defect and normal left ventricle. We considered whether primary biventricular repair was a safe alternative in all cases, even in the presence of interrupted aortic arch.

Methods: This was a retrospective analysis of patients who underwent primary biventricular repair consisting of a combination Norwood-type reconstruction of the aortic arch, baffle of the left ventricle to both semilunar roots, and conduit placement from the right ventricle to pulmonary arteries.

Results: Between January 1995 and January 2005, a total of 21 patients underwent primary biventricular repair at a median age of 5 days and a median weight of 3.0 kg. Aortic atresia was present in 7 and aortic stenosis in 14; 6 had interrupted aortic arch. All patients with aortic stenosis had annular diameters 3 mm or smaller. Median circulatory arrest time was 55 minutes, aortic crossclamp time was 56 minutes, and total support time was 99 minutes. In-hospital survival was 100%. Postoperative echocardiography in 19 patients demonstrated no significant outflow tract obstruction. Total stay was a median of 17 days. At midterm follow-up, there has been 1 late death, and reoperation has been necessary in 10 cases.

Conclusion: Primary biventricular repair is a safe alternative to staged repair in all cases of aortic hypoplasia with ventricular septal defect and normal left ventricle.

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Aortic atresia or severe aortic stenosis is usually associated with left ventricular hypoplasia. However, autopsy series reveal that in 4% to 7% of patients with aortic atresia, a ventricular septal defect (VSD) may exist, allowing an appropriate outflow for the ventricular chamber.[1-3] Through undefined embryologic mechanisms, this provides the left ventricle an opportunity for normal (or nearly normal) growth.

In the vast majority of cases of aortic atresia or severe aortic stenosis, the left ventricle does not form substantially, requiring the conversion to single-ventricle physiology for survival. Staged single-ventricle repair, though by now a standard operation, is associated with significant morbidity. Thus we have been motivated to preserve biventricular physiology when possible. Biventricular repair for selected patients with aortic atresia or severe aortic stenosis and a VSD is possible, thus averting staged single-ventricle repair. Early palliation for this disease was reported by Freedom and colleagues[4] in 1977; a Potts shunt and bilateral pulmonary arterial bands were used.[4] In 1983, Duffy and associates[5] reported a variation of the Freedom procedure, constructing a polytetrafluoroethylene shunt between the main pulmonary artery and the aorta.[5] Norwood and Stellin[6] used a conduit between the ventricular apex and descending thoracic aorta. Yasui and colleagues' reports[7,8] in

Abbreviations and Acronyms
VSD = ventricular septal defect

TABLE 1. Patient population (n = 21)

Demographic data	
Patients (No.)	21
Age at operation (d, median ± SD)	9.6 ± 13.2
Gestational age (wk, median ± SD)	37.5 ± 2.2
Sex (male/female)	11:10
Weight at operation (kg, median ± SD)	3.1 ± 0.8
Genetic syndrome (No.)	7
Noncardiac anomaly (No.)	6
Anatomy	
Aortic stenosis (No.)	14
Aortic atresia (No.)	7
Interrupted aortic arch (A/B)	1:5
Maximum aortic annular diameter (patients with aortic stenosis; mm)	3

1987 of 12 patients were the first to address a combined repair with a Rastelli-type reconstruction of the outflow tract and VSD closure with an interposition polytetrafluoroethylene graft reconstruction of the aortic arch. Seven patients, all with interrupted aortic arch, underwent primary biventricular repair with no operative mortality. Austin and coworkers[9] in 1989 reported a further modification; their 4 cases of primary repair resemble the operation most commonly used by our group today. This encompasses a Norwood-type reconstruction of the aortic arch with Damus-Kaye-Stansel anastomosis, followed by a Rastelli-type left ventricular baffle and right ventricular–pulmonary arterial conduit. During the past 15 years, further modifications by Bogers and associates,[10] Serraf and coworkers,[11] Bové and colleagues,[12-15] and others[16-18] have improved the understanding of operative issues on the management of these complex cases. This study summarizes our 10-year experience with the primary biventricular repair, examining midterm results.

Methods

Between January 1995 and January 2005, a total of 21 infants with either severe aortic stenosis (annulus <3.0 mm) or aortic atresia, a VSD, and a normal left ventricle underwent primary repair at the Children's Hospital of Philadelphia. We defined a normal left ventricle as an apex-reaching left ventricle with a mitral valve Z-score greater than −2.0. Six patients had interrupted aortic arch, 1 type A and 5 type B. Patients with other complex anatomic defects were excluded from our study. Median gestational age at birth was 37.5 weeks (range 32-40 weeks), with a median operative age of 9.6 days (range 1-59 days). Patient characteristics are reported in Table 1 and are notable for a balanced mix of male (n = 11) and female (n = 10) infants and a high frequency of either documented genetic syndromes (n = 7) or other noncardiac anomalies (n = 6). These genetic syndromes included either complete or microdeletions of 22q11 (n = 3), 4q32 (n = 1), trisomy 5q (n = 1), chromosome 9 partial inversion (n = 1), and *G6PD* deficiency (n = 1). All patients were supported preoperatively with prostaglandin E to maintain ductal patency and proceeded to operation as soon as was feasible in an elective fashion.

Operative Procedure

All patients were nasotracheally intubated and monitored with a radial arterial line. Surgery was tailored to individual anatomic variations, although the general procedure is as follows. Surgery was performed through a full midline sternotomy, and patients were cannulated in a standard fashion. Cardiopulmonary bypass was begun, and tapes were brought down around the branch pulmonary arteries. The patient's head was packed in ice, and the patient was cooled to 18°C through 15 minutes with alpha-stat blood gas management. During this period of cooling, the aortic arch was extensively mobilized and a homograft patch and a valved homograft conduit were thawed. The median conduit size was 12.0 mm (range 8-14 mm). Ramel tourniquets were placed loosely around the brachiocephalic vessels. A period of circulatory arrest began, cardioplegia (50 mL/kg) was infused through a side port of the aortic cannula, and cannulas were then removed. The patent foramen ovale was narrowed to between 2 and 4 mm. The ductus arteriosus was excised from the pulmonary bifurcation, and the distal aorta redundant ductal tissue was débrided. The inferior aspect of the aortic arch was then augmented with an arrowhead-shaped homograft patch, and the aortic and pulmonary roots were brought together in a Damus-Kaye-Stansel anastomosis. In cases of interrupted aortic arch, we usually anastomosed the lateral aspect of the right carotid and the medial aspect of the left subclavian artery to reconstruct the superior portion of the aortic arch. The inferior aspect of the aortic arch was reconstructed in an identical fashion. The right ventricular infundibulum was incised, and the ventricles were inspected. We generally did not resect the infundibular septum, although we enlarged the VSD in 11 cases. A Dacron polyester fabric patch baffled the left ventricular outflow tract to both the aortic and pulmonary valves. A valved homograft conduit was used to reconstruct the right ventricular outflow tract. Cannulas were replaced and perfusion begun, and the patient was warmed to 37°C through 23 minutes. All patients underwent a period of modified ultrafiltration after weaning from cardiopulmonary bypass. Total support time was a median of 99.0 minutes (range 77-141 minutes), with an aortic crossclamp time of 56.0 minutes (44-85 minutes) and a circulatory arrest time of 55.0 minutes (44-77 minutes). All patients underwent intraoperative transesophageal echocardiography for evaluation of the repair (Table 2).

Statistical Analysis

All statistical analyses were performed with SAS version 8.02 (SAS Institute, Inc, Cary, NC). Descriptive statistics were performed with PROC UNIVARIATE and PROC FREQ for continuous and categoric variables, respectively. Pearson correlation coefficients between continuous variables and measures of length of stay were calculated with the CORR procedure, and reported

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TABLE 2. Perioperative data

Operative data	
Total support time (min, median ± SD)	99.0 ± 24.0
Circulatory arrest time (min, median ± SD)	55.0 ± 8.0
Aortic crossclamp time (min, median ± SD)	56.0 ± 10.2
Conduit size (mm, median ± SD)	12.0 ± 1.7
Extracorporeal membrane oxygenation (No.)	0
Intensive care unit stay (d, median ± SD)	15.0 ± 30.0

P values test the null hypothesis that the correlation is zero. To assess differences in length of stay by categoric variables, such as the presence or absence of a genetic syndrome, unpaired *t* tests were performed as implemented in PROC T-TEST to test the null hypothesis that the difference between the two groups in mean length of stay was zero. The Satterthwaite method was used to calculate *P* values when there was evidence that the variances in the two groups were not equal.

Results

There were no operative or in-hospital deaths. No patients required extracorporeal membrane oxygenation support at any point during the hospital course. Median intensive care unit stay was 15.0 days (range 4-126 days). Complete follow-up was available for 16 of 21 patients, with a median follow-up of 34.2 months (range 0.5-81.9 months). Results are summarized in Table 3. Only gestational age predicted length of stay ($P = .02$). There were no important correlations between total support time ($P = .23$), circulatory arrest time ($P = .42$), and aortic crossclamp time ($P = .24$) and either length of stay or mortality. Interestingly, there was also no correlation between the presence of a genetic syndrome ($P = .24$) or noncardiac abnormality ($P = .24$) and length of stay.

There was 1 late death, from an unrelated event at an outside hospital, resulting in a 100% in-hospital survival and a 95.2% midterm survival. Significant complications occurred in 12 patients, with 10 reoperations. Eleven patients had baffle leaks, 4 of which required reoperation. There was 1 patch dehiscence at the tricuspid annulus and 3 moderate peripatch leaks. There were 7 trivial residual baffle leaks not requiring operative repair and 1 separate residual muscular VSD. Conduit intervention has been required in 11 patients to date, though all will require this at some point in the future. Conduit replacement has been required in 5 patients, all during separate hospital admissions, with no mortality and no significant morbidity. Conduit dilatation has been performed in 6 patients as a temporizing measure. Catheter balloon aortic arch dilatation has been required in 4 patients, although none have required further operative intervention. One patient required placement of an epicardial pacemaker for complete heart block. Valvular dysfunction occurred in 3 patients, with 1 requiring reoperation for severe tricuspid insufficiency. One additional patient had moderate tricuspid insufficiency, and 1 had moderate neoaortic insufficiency. No patients had left ventricular outflow tract obstruction. Median total stay was 17.0 days (range 8-154); 75% of patients had a total stay shorter than 24 days, and 90% had a total stay shorter than 49 days.

TABLE 3. Postoperative data

Residual baffle leak or VSD (No.)	11
Baffle leak necessitating operation	4
Arch balloon dilatation (No.)	4
Conduit revision (No.)	5
Conduit dilatation	6
Epicardial pacemaker	1
Tricuspid insufficiency	2
Neoaortic insufficiency	1
Total reoperations (No.)	10
Late deaths (No.)	1
Length of stay to discharge (d, median ± SD)	17 ± 31.7

Conclusions

This retrospective analysis of 21 patients undergoing the primary biventricular repair of aortic atresia or aortic stenosis between 1995 and 2005 shows acceptable midterm results. Primary biventricular repair is now our procedure of choice for patients with either aortic atresia or severe aortic stenosis with a normal left ventricle and VSD. The presence of interrupted aortic arch does not influence our decision; in fact, there are no instances in which we preferentially undertake a staged repair. Residual or developing subaortic stenosis has not been a problem in our patient population. Reinterventions are a necessity; all patients will eventually return for conduit revisions.

The most troublesome problem was baffle leak, which necessitated reoperation in 4 cases. This consistent problem emphasizes the need for close attention to this technical aspect of the intracardiac repair. We could not identify a consistent location of these defects, nor was there a consistent correlation with VSD enlargement. We therefore recommend careful examination of the patch before completion of the proximal conduit anastomosis. Of note, all our patches were completed in a running fashion; we therefore cannot exclude the possibility that an interrupted approach may be more secure in this situation.

Considerations for repair are determined by the spectrum of disease. One issue that remains is the consideration of single-stage versus staged repair. Ohye and coworkers[13] compared primary versus staged repair in 20 patients, 11 of whom underwent a single-stage repair. There were no significant differences in morbidity or mortality between the groups with staged and unstaged repair, suggesting no clear superiority of either staged or primary repair. A recent report by Pearl and associates[19] on a staged approach in 8 patients had no mortality, suggesting that this continues to be safe approach. Arguments for primary repair include the possible reduction in number of operations, early relief of relative cyanosis, and reduction in ventricular volume load. Proponents of a staged approach argue that a reduction in neonatal ischemic myocardium or removal of a neonatal ventriculotomy may improve outcome. Our data set cannot address the superiority of primary versus staged biventricular repair, because all patients underwent primary repair. Similarly, we cannot address the important question, especially in cases of marginal left ventricular size, of whether a staged single-ventricle palliation leading to a Fontan procedure would be superior to primary biventricular repair.

This study has limitations and cannot address a number of important questions. For example, who is suitable for single-ventricle versus biventricular repair? We did not examine this issue because all ventricles were deemed adequate, with an apex-forming left ventricle and a mitral valve Z-score greater than -2.0. Although we cannot support the statement with data, the good intermediate outcome of our patients suggests that outcome may not be improved by a staged repair. Overall, we suggest that primary biventricular repair of aortic atresia or severe aortic stenosis with a good left ventricle is safe at intermediate follow-up and should be considered for this complex set of patients.

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